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CASE REPORT
Hand

Idiopathic Calcinosis Cutis of the Hand

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Summary: Calcinosis cutis is a disease process characterized by calcified lesions in the skin. Although there are five subtypes of calcinosis, idiopathic calcinosis cutis is a rare disease process with no clear etiology. It has been described in many parts of the body; however, there are only five reported cases specifically involving the hands. We describe the presentation and successful treatment of a case of idiopathic calcinosis cutis in a 65-year-old man with lesions on his bilateral hands. We believe that surgical excision of symptomatic lesions is a safe and effective treatment for idiopathic calcinosis cutis of the hands. *(Plast Reconstr Surg Glob Open 2024; 12:e5849; doi: 10.1097/GOX.0000000000005849; Published online 24 May 2024.)*

Calcinosis cutis refers to the deposition of insoluble calcium salts in the skin and subcutaneous tissue.[1] There are five different subtypes: dystrophic, metastatic, iatrogenic, calciphylaxis, and idiopathic. Dystrophic calcinosis cutis, the most common form, is caused by tissue damage and commonly associated with autoimmune connective tissue diseases, such as systemic sclerosis, dermatomyositis, and systemic lupus erythematosus.[1] Metastatic calcinosis cutis results from defective metabolism of calcium and phosphate. Iatrogenic calcinosis cutis occurs from treatment with intravenous calcium compounds and calcium chloride electrode paste used in electroencephalography. Calciphylaxis is characterized by the calcification of small- and medium-sized blood vessels in the dermis or subcutaneous fat, leading to ischemia, necrosis, and ulceration of the skin.[1]

Idiopathic calcinosis cutis is a rarer form of this disease process.[2–6] It occurs without any underlying tissue damage or defects in metabolism and can present as papules, nodules, or plaques that may be ulcerated or with white chalky discharge.[2–6] The pathophysiology remains unknown but may be related to a defect in the metabolism of gamma carboxy glutamic acid.[7] There are four main types of idiopathic calcinosis cutis: scrotal calcinosis, consisting of hard, yellowish, asymptomatic nodules on the scrotal skin[7]; subepidermal calcified nodules, presenting as an asymptomatic, solitary, white-yellow, hard papule commonly located on the head and neck region of male children[8]; tumoral calcinosis with firm, painless, tumor-like masses around the joints[9]; and milia-like calcinosis cutis, involving multiple small, firm, white- or skin-colored papules commonly found in the hands and feet.[10] To date, only five cases of idiopathic calcinosis cutis have been described in the literature.[2–6] As such, we present this rare case involving surgical management of idiopathic calcinosis cutis in the hand.

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CASE DISCUSSION

A 65-year-old right-handed man presented to the clinic with painful bumps on his bilateral hands. He first noticed these lesions approximately 8 years prior. He underwent incisional biopsy of the lesion on his right middle finger about 1 year before presentation, but the pathology report was not available to review. The patient noted that the lesion regrew.

The patient had previously been seen by a rheumatologist. The working diagnosis was gout, although no changes were observed after prescribing allopurinol, and uric acid levels were not elevated. Allopurinol was discontinued, and rheumatologic workup revealed no signs of autoimmune disease or renal dysfunction.

The patient's chief complaint was pain associated with the lesions. He was also concerned about obtaining a definitive diagnosis for these lesions on his hand. Plain films of the hand demonstrated multiple discrete calcified lesions (Figs. 1 and 2). The patient also had medication-controlled type 2 diabetes and was on a beta-blocker for hypertension. Surgical history included bilateral total knee arthroplasty.

To relieve the patient's symptoms and establish a diagnosis, excisional biopsy of four masses was performed on his right hand. Intraoperatively, the masses were noted to be ulcerated, poorly encapsulated, and calcified. Each was removed with an elliptical incision before the wound was irrigated and explored until the bulk of each lesion had

Disclosure statements are at the end of this article, following the correspondence information.

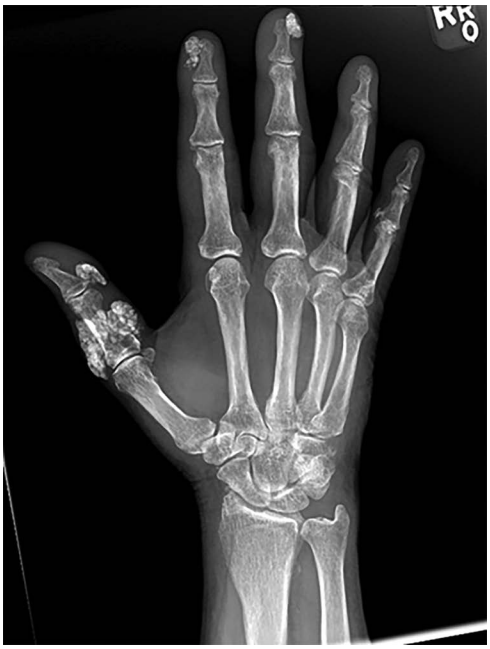

Fig. 1. Plain film of the patient's right hand demonstrating calcified lesions diagnosed as calcinosis cutis of the thumb, index finger, long finger, and small finger. The painful lesion on the thumb is noted to be volar to the base of the thumb's distal phalanx.

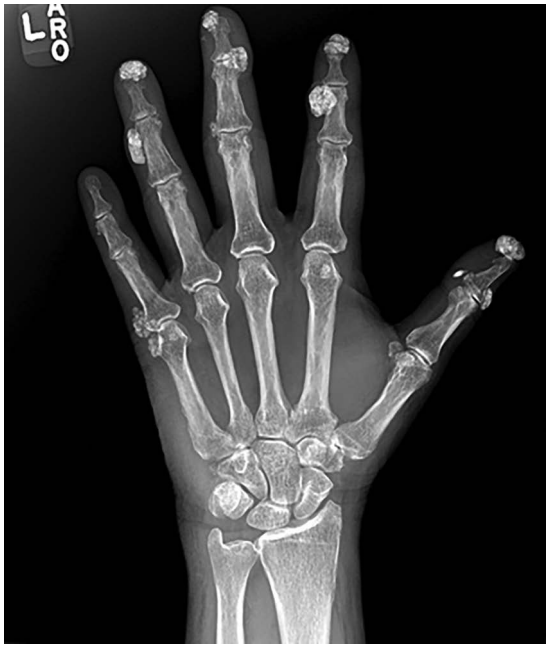

Fig. 2. Plain film of the patient's left hand demonstrating calcified lesions diagnosed as calcinosis cutis of all digits, although none of these lesions were painful.

been removed. As the distal thumb lesion involved the radial digital nerve, primary nerve repair was completed after mass excision. Histopathology was consistent with nodular calcinosis cutis.

Postoperatively, the patient healed well with significant pain reduction. After 1 year, there was no evidence of recurrence. As the lesions of his left hand were stable and asymptomatic, he elected to forgo treatment of his left hand. Given his lack of any contributory clinical history, he was diagnosed with idiopathic calcinosis cutis.

DISCUSSION

The diagnosis of idiopathic calcinosis cutis was made for this patient because there was no previous trauma; evidence of an inflammatory process; history of parenteral therapy; or abnormalities in calcium, phosphorous, or renal function. This case does not fall into the previously described four subtypes of idiopathic calcinosis cutis. Although milia-like calcinosis commonly occurs in the hand, this subtype typically presents as papules resembling milia, which was inconsistent with the clinical picture of our patient.[10] Of the five reported cases of idiopathic calcinosis cutis in the literature, three cases were isolated to the hands, whereas the other two included both elbows and lower extremities.[2–6] (Table 1).

The clinical presentations of calcinosis cutis and tophaceous gout share some similarities—they both, for example, may present with firm, ulcerated lesions with chalky extrusion of calcium. However, a calcinosis cutis lesion seems basophilic on hematoxylin and eosin stain and is typically confirmed with von Kossa or alizarin red stains, which specifically highlight calcium.[11] This contrasts the typical needle-like negatively birefringent monosodium urate crystals of gout.

There is no standard therapy for calcinosis cutis, but several treatments have been shown to be beneficial to varying degrees. These include diltiazem, warfarin, bisphosphonates, minocycline, ceftriaxone, intravenous immunoglobulin, surgical excision, carbon dioxide laser, extracorporeal shock wave lithotripsy, curettage, and probenecid.[6]

Surgical excision, as demonstrated in the presented case, is indicated for pain, recurrent infection, ulceration, functional impairment, and localized lesions. Our patient's symptomatic calcification in his right thumb was the primary indication for surgery, in addition to obtaining a histopathologic diagnosis. Surgical risks in the hand include damage to the digital neurovascular bundles and wound complications, with the caveat that excised lesions may recur. In our experience, these pitfalls can be avoided with complete excision, careful dissection, and identification of sensitive anatomy. Surgical excision of idiopathic calcinosis cutis of the hand is an effective and definitive treatment for these lesions, with no reported recurrences in the literature.[2–6] Medical treatments, including colchicine and diltiazem, however, seem to be ineffective for idiopathic calcinosis cutis of the hand.[2,3]

Patients with localized lesions are generally good candidates for surgical excision. This, however, must be balanced with the risk of damage to deep structures if neuromuscular involvement is suspected. In this patient's

Table 1. Review of Reported Idiopathic Calcinosis Cutis Cases

Author (Year Published)	Age, y	M/F	Location	Other Affected Areas	Treatment	Outcome
Terranova et al (2005)[6]	43	M	Bilateral hands	None	Not described	Not described
Guermazi et al (2006)[2]	66	F	Bilateral hands	Bilateral elbows, right knee, left foot	Colchicine ×3 y (no effect); surgical excision	Not described
Sato et al (2007)[5]	35	M	Left long finger	None	Surgical excision	No recurrence after 8 y
Prabhu et al (2014)[4]	59	F	Right thumb	Right elbow, left great toe	Local wound care	Not described
Meena et al (2020)[3]	52	F	Bilateral hands	None	Oral diltiazem	No improvement after oral diltiazem
Our case	65	M	Bilateral hands	None	Surgical excision	No recurrence to date

M, male; F, female.

case, primary nerve repair was possible despite partial radial digital nerve resection with nodule excision. Before his procedure, we counseled this patient that neurovascular structures may be compromised, and complete debulking may be abandoned intraoperatively if those risks were deemed too great. In patients with particularly large lesions or extensive involvement of critical structures, more conservative debulking may be offered with the understanding that the risk of recurrence may be higher in these cases. Although the relative rarity of the idiopathic subtype limits our understanding of its incidence of recurrence, multiple studies demonstrate high recurrence rates in other subtypes of the disease.[12]

Although idiopathic calcinosis cutis of the hand is a rare diagnosis, it should be considered when a patient presents with otherwise unexplained calcifications. The literature clearly demonstrates that calcinosis cutis is often associated with other disease processes, though there are cases where the underlying etiology is unclear. We found that surgical excision of symptomatic calcinosis cutis lesions of the hand is safe and effective.

RECOMMENDATIONS

When encountering patients like ours, the primary recommendation is to determine the etiology of the nodule. Idiopathic calcinosis cutis is a diagnosis of exclusion.[1,2] This involves referral to dermatology and rheumatology to investigate any potential underlying medical conditions.

Depending on the location of the lesion, surgical excision can be deforming and debilitating, especially if one is not cognizant of the (likely distorted) relevant surgical anatomy. Although these lesions do not pose any inherent danger (such as risk of metastasis), pain and infection from open sores may develop. It is for these reasons we recommend surgical excision of symptomatic lesions.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

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